

FRG1 siRNA (h): sc-62350

BACKGROUND

FRG1 is a 258 amino acid nuclear protein encoded by the human gene FRG1. The FRG1 protein is thought to be involved in pre-messenger RNA splicing. FRG1 plays a role in processing pre-rRNA, assembling rRNA into ribosomal subunits and may also be involved in pre-mRNA splicing. Facioscapulohumeral muscular dystrophy (FSHD) is a disease state associated with internal deletions among the tandem array of D4Z4 repeats on chromosome 4q35, a subtelomere region of chromosome 4 that contains the FRG1 gene. The muscle degeneration that is common in patients with FSHD results from increased expression of genes proximal to the deletion, including FRG1. In addition to muscle degeneration, most FSHD patients also develop abnormalities of the retinal vasculature. FRG1 is expressed in adult and fetal muscle, lymphocytes and placenta. It can be localized to nuclear Cajal bodies or speckles.

REFERENCES

1. van Deutekom, J.C., et al. 1996. Identification of the first gene (FRG1) from the FSHD region on human chromosome 4q35. *Hum. Mol. Genet.* 5: 581-590.
2. Grewal, P.K., et al. 1997. The mouse homolog of FRG, a candidate gene for FSHD, maps proximal to the myodystrophy mutation on chromosome 8. *Mamm. Genome* 8: 394-398.
3. Grewal, P.K., et al. 1998. FRG, a gene in the FSH muscular dystrophy region on human chromosome 4q35, is highly conserved in vertebrates and invertebrates. *Gene* 216: 13-19.
4. Grewal, P.K., et al. 1999. Recent amplification of the human FRG1 gene during primate evolution. *Gene* 227: 79-88.
5. Tam, R., et al. 2004. The 4q subtelomere harboring the FSHD locus is specifically anchored with peripheral heterochromatin unlike most human telomeres. *J. Cell Biol.* 167: 269-279.
6. van Koningsbruggen, S., et al. 2004. FRG1P is localised in the nucleolus, Cajal bodies, and speckles. *J. Med. Genet.* 41: e46.
7. Gabellini, D., et al. 2006. Facioscapulohumeral muscular dystrophy in mice overexpressing FRG1. *Nature* 439: 973-977.
8. Osborne, R.J., et al. 2007. Expression profile of FSHD supports a link between retinal vasculopathy and muscular dystrophy. *Neurology* 68: 569-577.

CHROMOSOMAL LOCATION

Genetic locus: FRG1 (human) mapping to 4q35.2.

PRODUCT

FRG1 siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see FRG1 shRNA Plasmid (h): sc-62350-SH and FRG1 shRNA (h) Lentiviral Particles: sc-62350-V as alternate gene silencing products.

For independent verification of FRG1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-62350A, sc-62350B and sc-62350C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

FRG1 siRNA (h) is recommended for the inhibition of FRG1 expression in human cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

GENE EXPRESSION MONITORING

FRG1 (C-5): sc-377040 is recommended as a control antibody for monitoring of FRG1 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor FRG1 gene expression knockdown using RT-PCR Primer: FRG1 (h)-PR: sc-62350-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.